

**PRIMARY CARDIAC BURKITT LYMPHOMA PRESENTING AS A RIGHT ATRIAL MASS: CASE REPORT AND REVIEW OF THE LITERATURE.****Amina Samih<sup>\*1</sup>, Hanae Bouhdadi<sup>2</sup>, Sabine Derqaoui<sup>3</sup>, Bernoussi Zakia<sup>3</sup>, Mohammed Laaroussi<sup>2</sup> and Mohammed Cherti<sup>1</sup>**<sup>1</sup>Department of Cardiology B, Maternity Hospital IBN Sina, Rabat - Morocco.<sup>2</sup>Department of Cardiovascular surgery, IBN Sina Hospital, Rabat - Morocco.<sup>3</sup>Department of Pathology, IBN Sina Hospital, Rabat - Morocco.**\*Corresponding Author: Amina Samih**

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**ABSTRACT**

Primary cardiac lymphoma (PCL) is an uncommon and very rare entity, accounting only for 1% of extra nodal lymphoma with heart involvement. Infections and thrombi account for most non tumoral intracardiac masses. Primary tumors of the heart are rare. Some 75% of the tumors are benign, 25% malignant. Because of the low case numbers, there is an insufficient evidence base to determine the optimal treatment, particularly for malignant tumors. **Case Summary:** We report the case of a young woman who complained of breathlessness. A transthoracic echocardiography revealed a large mobile homogeneous mass in the apical 4 chamber view, which appeared to be arising from the interatrial septum in the right atrium with tricuspid valve stenosis. The mass impaired right ventricular. Urgent surgery for debulking was indicated. Pathology was a burkitt lymphoma (BL). **Discussion:** BL causing intracardiac mass is rare. The cardiac symptoms are frequently non-specific and can often be overlooked or underappreciated in the presence of the impressive extra-cardiac disease. This observation is interesting to emphasize the role of early diagnosis that improves the prognosis despite a non-specific clinical presentation.

**KEYWORDS:** Non hodgking lymphoma • burkitt's lymphoma • Primary cardiac lymphoma • histology • Surgical debulking.

**INTRODUCTION**

PCL is a rare extra nodal lymphoma involving only the heart with or without pericardial involvement. It accounts only for 1,6% of malignant cardiac tumors which represent 30% of cardiac tumors and less than 1% of extra nodal lymphomas. Burkitt Lymphoma constitutes less than 1% of all non-Hodgkin lymphomas. Its frequency is underestimated, because 20% of cases were diagnosed at autopsy.

Its non-specific presentation leads usually to delayed diagnosis and poor prognosis.

**CASE PRESENTATION**

We present the case of a 35-year-old woman who presented to the emergency department for dyspnea, with a history of three weeks of shortness of breath associated to chest pain exacerbated by coughing and deep inspiration. Two days before her admission, the patient reported a worsening dyspnea from NYHA class II to NYHA class III without orthopnea. The patient had no history of smoking or another substance abuse. She had

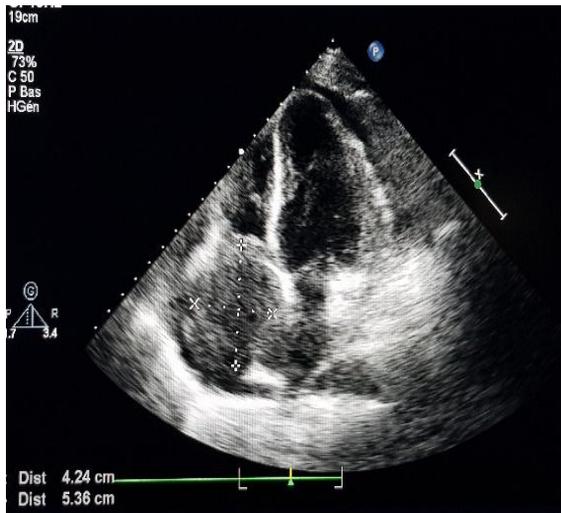
no history of diabetes or hypertension. No special medical background was reported.

In physical examination, body temperature was 37,9C. She was in poor general condition, polypneic at 25 cycles/min with a heart rate at 100 pulse/min. Blood pressure was 110/75 mmHg. She had jugular vein distention, a superior vena cava syndrome and bilateral leg oedema.

A 12-lead electrocardiogram showed sinus rhythm with a complete right bundle block. Chest X ray revealed a cardiomegaly and pleural effusion.

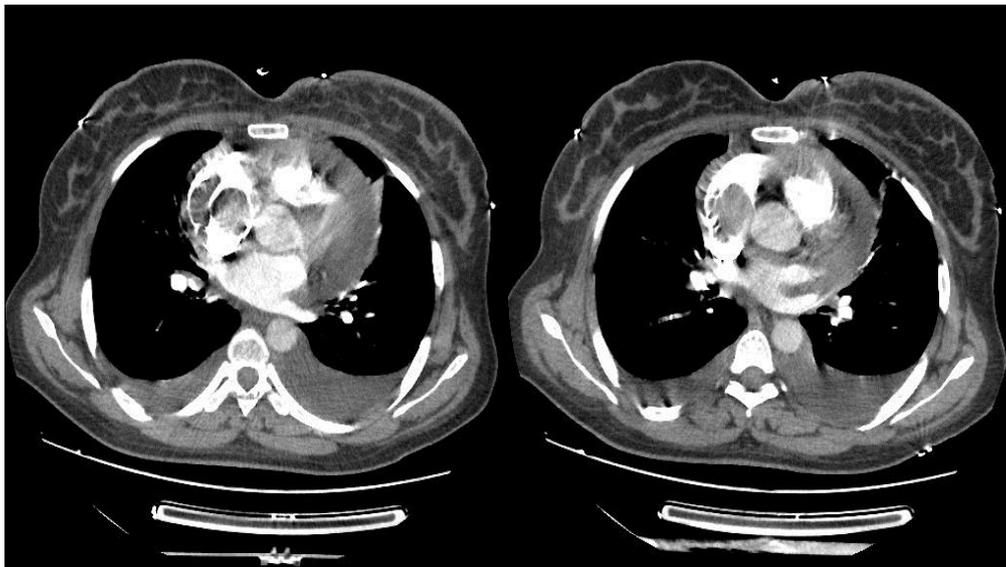
Transthoracic echocardiography showed mobile homogeneous mass measuring 5,36cmx4,24cm in the apical 4 chamber view (Figure 1), which appeared to be arising from the interatrial septum in the right atrium with tricuspid valve stenosis. The mass impaired right ventricular hemodynamics. A large pericardial effusion (20mm) was noted. However, the effusion was not haemodynamically significant. There was no ventricular or atrial collapse, no apparent transmitral respiratory variation. There was no extension to the superior vena

cava or inferior vena cava. The ejection fraction was preserved.



**Figure 1: Echocardiography showing a mass arising from the interatrial septum in the right atrium.**

Thoracic CT scan does not show a pulmonary embolism (Figure 2A and Figure 2B).



**Figure 2A: and figure 2B: Thoracic CT scan eliminates pulmonary embolism.**

To investigate for other tumours or metastases, abdominal CT scan was done but showed no other tumour.

The biological assessment showed anemia at 10.5 g/dl of hemoglobin, 12000/mm<sup>3</sup> hyperleucocytosis and 150 mg/l of reactive protein C and raised serum creatinine 20 mg/l.

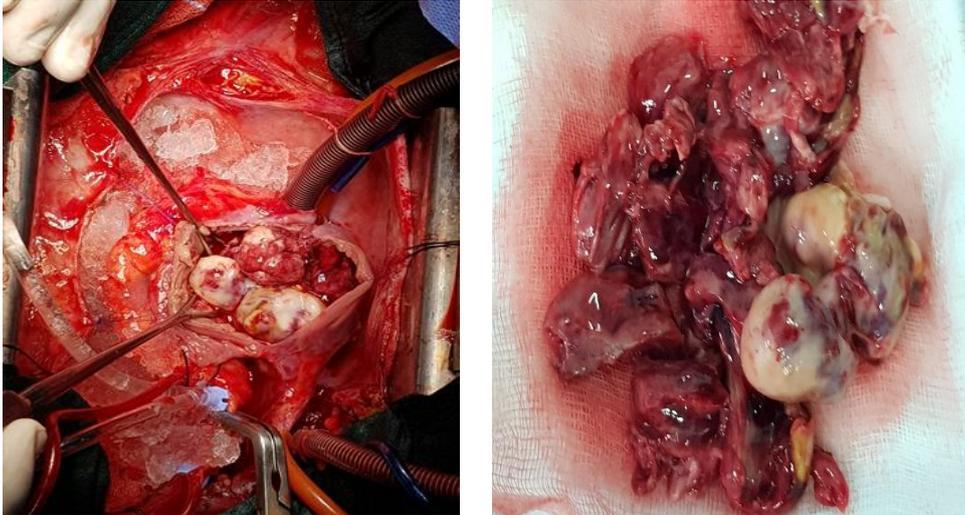
Arterial blood gas showed severe acidosis with reduced pH 7.22, reduced bicarbonate 12mmol/L and markedly increased blood lactate 13 mmol/L.

Urgent surgery for debulking was indicated.

We proceeded by a median sternotomy. After the opening of the pericardium and aspiration of pericardial

effusion. A routine cardiopulmonary bypass between an aortic cannula and two laced vena cava (we used a curved superior vena cava cannula), after aortic cross-clamping and adequate myocardial protection, the right atrium was opened: a globular, heterogenous, yellowish tumour with unclear was seen (Figure 3).

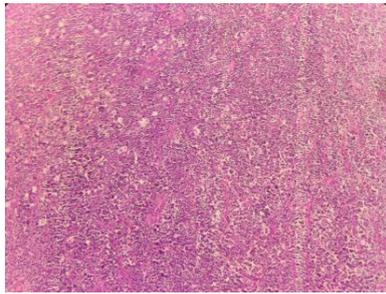
Debulking was performed until the tricuspid valve became visible. The annulus appeared dilated so tricuspid valve repair was done using an annuloplasty ring.



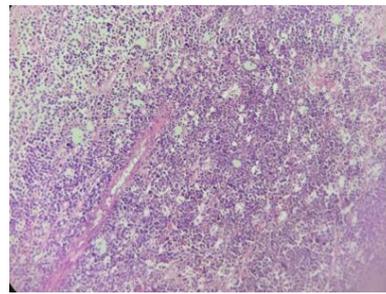
**Figure 3: Intraoperative pictures of the tumour.**

The histopathology and immunohistochemistry examination of the resected specimen demonstrated, on gross, removed cardiac fragment's diameter ranged from 1 cm to 5 cm, presenting as grey and homogenous masses. Hematoxylin-eosin stained sections revealed a diffuse pattern proliferation of intermediate-to-large lymphoid cells with irregular nuclei, small centrally located nucleoli and a basophilic cytoplasm. A Starry sky

appearance was present due to the numerous tangible body macrophages (Figure 4A and figure 4B). The proliferation shows multiple mitotic figures (Figure 5). On immunohistochemistry, the neoplastic cells expressed CD20, CD10 and Bcl 6. They were negative for Bcl 2 and CD5. The Ki67 proliferation's rate was 100% (Figure 6).

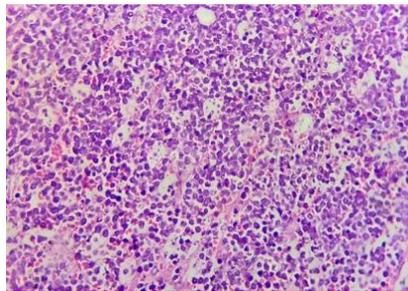


**4A**

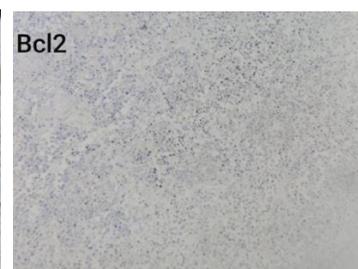
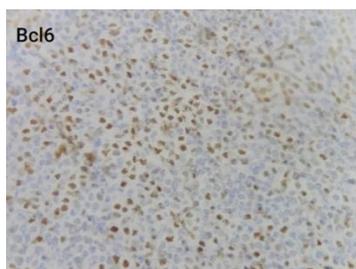
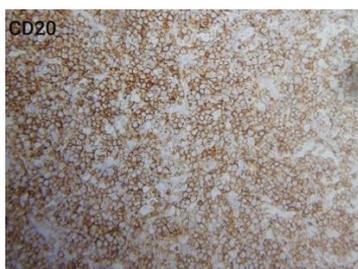


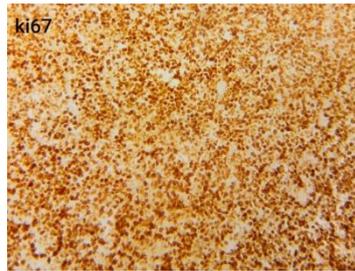
**4B**

**Figure 4A: and figure 4B: Burkitt's lymphoma starry sky appearance (HE: x100 (1) x 200 (2)).**



**Figure 5: Multiple mitotic figures (HE x400).**





**Figure 6: Immunocytochemistry of Burkitt's lymphoma: CD20+, Bcl6+, CD10+, Bcl2-, Ki-67.**

After surgery, the patient was admitted to the ICU with hemodynamic instability requiring high doses of catecholamines. Unfortunately, the patient died within the 48 postoperative hours due to a severe right ventricular dysfunction complicated by a multiple organ failure.

## DISCUSSION

Primary cardiac tumors are uncommon, estimated to be 100-1000 times less prevalent than metastases.<sup>[1]</sup>

About 90% of all primary cardiac tumors are benign and include myxomas, papillary fibroelastomas, fibromas, lipomas, angiomas, and rhabdomyomas. Sarcomas are the most common malignant primary cardiac tumors, accounting for 95% of cases, and mesotheliomas and lymphomas account for the rest.<sup>[2]</sup>

Primary cardiac lymphoma (PCL) is extra-nodal lymphoma involving only the heart and/or the pericardium.<sup>[3]</sup> It is rare, accounting only for 1.6% of malignant cardiac tumours which only account for 30% of cardiac tumours.<sup>[4]</sup>

BL is a highly aggressive subset of B cell NHL, characterized by a very high proliferation rate. It is an uncommon disease in adults, accounting for less than 1% of all NHL.<sup>[5]</sup>

HIV patients have a 200-fold increased risk of developing NHL, with 25% to 30% of these being BL or Burkitt-like lymphoma.<sup>[6]</sup>

PCL has a propensity to involve the superior vena cava (SVC) and the right side of the heart, which are the areas that receive lymph drainage from the thoracic duct. It is hypothesized that PCL could be merely a manifestation of occult systemic lymphoma (spreading to the heart by the thoracic duct) instead of a primarily cardiac issue.<sup>[7]</sup> Regardless of the origin, an intracardiac mass caused by BL is exceedingly rare. To the best of our knowledge, there are only 22 reported cases in the English literature (Table 1).

Among the 23 cases (including present case), all 3 types of BL were represented (10/23 (43%) sporadic, 4/23 (17%) endemic, 9/23 (40%) immunodeficiency-associated). Most were men (17/23 (74%)). The age ranged from 4 to 78 years, with a median of 38 years, much younger than

the median age for PCL in adults, which is approximately 60 years.<sup>[7]</sup> The reason for this predilection for intracardiac involvement in younger patients remains unclear. Common presenting symptoms include dyspnea, palpitation, dizziness, and B-symptoms, which are consistent with prior studies on cardiac lymphoma.<sup>[6,7]</sup> There were 9/23 (39%) HIV-positive cases, 8/23 (35%) HIV-negative cases, and HIV status was not reported in 6/23 (26%). The disease was right-sided in 17/23 (74%) cases.

Patients received standard chemotherapy regimens such as EPOCH (etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin) or CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) with or without rituximab, Hyper-CVAD (cyclophosphamide, vincristine, doxorubicin, and dexamethasone), and CODOX-M. A total of 11/23 (48%) patients died.

Most of these patients died within a few days of diagnosis, 10/11 (90%), and 1 died 6 weeks after diagnosis. BL was diagnosed at autopsy in 2 cases. The longest surviving patient was reported to be alive at 36 months after diagnosis. It is well known that BL is a highly aggressive tumor, but in the presence of cardiac involvement, it seems to lead to even more complications and worse outcomes.

**Table 1: Burkitt-like or highly likely Burkitt;\*\* small non-cleaved cell type. Ara-C – cytarabine; BLE – bilateral lower extremity; CHOP – cyclophosphamide, doxorubicin, vincristine, prednisone; CODOX-M – cyclophosphamide, vincristine, doxorubicin, and methotrexate; COP – cyclophosphamide, vincristine, prednisone; COPADM – cyclophosphamide, vincristine, prednisone, doxorubicin, methotrexate; CP – chest pain; CVAD – cyclophosphamide, vincristine, doxorubicin, dexamethasone; CYM – Ara-C, methotrexate; D – deceased; D/C – discharged; EPOCH – etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin; F – fever; HA – headache; HD – high-dose; LA – left atrium; LN – lymph node; LTF – lost to follow-up; LV – left ventricle; m – month(s); M – man; MTX – methotrexate; N/A – information not available; Neg – negative; + – positive; R – remission; R-CHOP – rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone; R-EPOCH – rituximab, etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin; RA – right atrium; RV – right ventricle; SOB – shortness of breath; TV – tricuspid valve; W – woman; wks – weeks; WL – weight loss; yrs – years.**

Year	Age	Sex	HIV	Pertinent symptoms	Intracardiac mass location	Diagnostic procedure	Treatment	Outcome	Reference
1975	12	M	N/A	SOB, anorexia, palpitation	RA	Autopsy	No treatment	D (days)	Cole <sup>[10]</sup>
1990*	35	M	+	SOB, WL, orthopnea	RV	Pericardiotomy	Vincristine	D (days)	Helfand <sup>[11]</sup>
1992	29	M	N/A	SOB, WL, F	RA, LA	Laparotomy	N/A	D (days)	Zyssman <sup>[12]</sup>
1992**	13	M	N/A	F, cough	LA	Mediastinoscopic biopsy	N/A	R	Moore <sup>[13]</sup>
1992**	55	W	N/A	Syncope	RA	Autopsy	No treatment	D (days)	Bestetti <sup>[14]</sup>
1998	47	M	+	SOB	TV, RV, RA, LA	Cytology of pericardial fluid	EPOCH, bleomycin	R (3+ yrs)	Brinkman <sup>[15]</sup>
2000	78	W	N/A	SOB	RA	Thoracotomy	N/A	D (days)	Carfagna <sup>[16]</sup>
2004	10	M	Neg	SOB, cough, fatigue	RA, inter-atrial septum, pulmonary infundibulum	Thoracotomy	Patient was in a study: 1 x COP, COPADM, CYM	R (3+ yrs)	Chalabreysse <sup>[17]</sup>
2005	9	M	N/A	SOB	RA	N/A	N/A	R (4 wks)	Ahmad <sup>[18]</sup>
2006*	70	M	Neg	SOB	RA	Endomyo-cardial transvenous biopsy	N/A	N/A	De Filippo <sup>[19]</sup>
2006	4	M	+	SOB, orthopnea	RA	Median sternotomy	N/A	D (days)	Singh <sup>[20]</sup>
2007	41	W	+	BLE weakness, lower back pain	LV	Vertebral mass biopsy	EPOCH	R	Mendiolaza <sup>[21]</sup>
2007	52	M	+	SOB, CP, night sweats	RA, LA	N/A	R-CHOP	R	Poh <sup>[22]</sup>
2008	61	W	Neg	SOB, palpitation	RV	Thoracotomy	Hyper-CVAD	D (days)	Stefani <sup>[23]</sup>
2009	33	M	Neg	SOB	RA	Thoracotomy	N/A	N/A	Peng <sup>[24]</sup>
2009	67	M	Neg	Syncope	RA, LA	Resection	CODOX-M, Ara-C	R (12 m)	Santini <sup>[25]</sup>
2009*	74	M	Neg	SOB	RA	Intracardiac mass biopsy	CHOP	D (6 wks)	Legault <sup>[26]</sup>
2010*	9	M	Neg	SOB, cough, palpitations	RA	Fine needle cervical node biopsy	CHOP	D/C after 3 wks, LTF	Mocumbi <sup>[27]</sup>
2014*	45	M	+	F, WL, night sweats	LV	Liver biopsy	R-EPOCH	D (days)	Bush <sup>[6]</sup>
2014	26	F	+	SOB, leg edema, fatigue	RA	Laminectomy	CHOP	D (days)	Basavaraj <sup>[28]</sup>
2015	38	M	+	SOB, palpitations	RA	Open sternotomy	R-EPOCH	R	Lazkani <sup>[29]</sup>
2016	27	M	+	Palpitations, WL, Dizziness	RA, RV	Liver Biopsy	CODOX-M/IVAC	R	Onyee <sup>[30]</sup>

BL has a rapid doubling time, and intracardiac involvement can become life-threatening very quickly, as seen in our patient.

Surgical resection of PCL is difficult and have not shown any benefit. It should be avoided especially when diagnosis can be released by less invasive technics such as endomyocardial biopsy, or pericardial fluid sampling. However, surgery should be reserved for patients with life-threatening compromising hemodynamic state, as was the case of our patient.

Early systemic treatment appears to be the only chance for cure. Chemotherapy remains the preferred initial treatment; the CHOP protocol is most often employed. Complete remission has been achieved in less than 60% of cases.<sup>[31]</sup>

## CONCLUSION

Burkitt lymphoma causing intracardiac mass is rare. The cardiac symptoms are frequently non-specific and can often be overlooked or underappreciated in the presence of the impressive extra-cardiac disease. The cardiac tumour can progress and become life-threatening very quickly. Any delay in diagnosis or treatment can decrease the chances of survival.

The prognosis of cardiac Burkitt lymphoma remains very reserved despite the therapeutic progress due to its aggressive character.

**Consent:** The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient.

**Author's Contributions:** All authors have read and agreed to the final version of this manuscript and have equally contributed to its content and to the management of the manuscript.

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